Case report

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Exuberant Proliferating Trichilemmal Tumor in a young person

Tumor triquilemal proliferante exuberante em jovem

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ABSTRACT

Proliferating Trichilemmal Tumor (PTT) is an uncommon benign neoplasm that usually affects adult women over 60 years old, preferably as a solitary nodule in the scalp. We report a case of PTT in a young woman presenting exuberant, asymptomatic scalp tumors 7 years ago, which recurred after surgical exeresis. The histopathology confirmed the diagnosis, and surgical excisions and clinical follow-up were performed. PTT appears to originate from the wall of trichilemmal cysts, from the isthmus of the hair follicle's outer root sheath. Usually, it has benign biological potential with a low risk of malignancy and high recurrence. The therapy of choice is surgery.

Keywords: Cysts; Neoplasms; Scalp

RESUMO

Tumor triquilemal proliferante (TTP) é uma neoplasia benigna incomum que acomete, habitualmente, mulheres adultas acima de 60 anos, preferencialmente como nódulo solitário no couro cabeludo. Relata-se um caso de TTP em mulher jovem, apresentando exuberantes tumorações assintomáticas no couro cabeludo, há sete anos, que recorreram após exéreses cirúrgicas. O histopatológico confirmou o diagnóstico, tendo sido realizadas retiradas cirúrgicas e acompanhamento clínico. O TTP parece originar-se da parede dos cistos triquilemais, provenientes do istmo da bainha radicular externa do folículo piloso. Geralmente, apresenta potencial biológico benigno, com baixo risco de malignização e elevada recorrência. A terapia de escolha é a cirurgia.

Palavras-chave: *Cistos; Couro Cabeludo; Neoplasias*

INTRODUCTION

Proliferating trichilemmal tumor (PTT) is an epidermal adnexal neoplasia formed by several cysts containing squamous epithelium with trichilemmal keratinization, in other words, the abrupt transition of a nucleated epithelial cells to an anucleated in the absence of a granular layer.[1-3] PTT usually appears as a dermal nodule or solitary subcutaneous scalp of elderly women.[1,3-13] It was first described by Wilson-Jones in 1966, who named it a proliferating epidermoid cyst due to its clinical and histological resemblance to squamous cell carcinoma.[1,3, 5,9-13] Since then, PTT has received several other names, such as scalp pilaris tumor, proliferating trichilemmal cyst, trichilemic pillar tumor, invasive pilomatrixoma, hydatid keratinous cyst, trichoclamidocarcinoma, giant hair matrix tumor, hairy scalp tumor, and others.[1,3,6] It exhibits the ability to grow rapidly over

a given period of time, but the evolution to malignant PTT is uncommon.[5,7,11,12] Therefore, it presents heterogeneous histological behaviors ranging from benign, locally aggressive growth with chances of causing invasions in the underlying tissue layers up to that time. with local recurrence metastatic potential and regional lymph node metastasis.[3,10,11,12] PTT is a rare tumor that corresponds to 0.1% of benign cutaneous tumors and may form from a pre-existing or concomitant trichilemmal cyst or as an isolated lesion.[5,11,12]

CLINICAL CASE

We report the history of a 24-year-old female patient from Uberlândia (MG), treated at the dermatology department of the Clinical Hospital of the Federal University of Uberlândia, complaining of painless nodules in scalp for seven years ago. It refers to progressive growth in recent months, followed by ulceration with output of bloody discharge. Reports recurrence of previously excised scalp lesions. She denies having suffered any kind of trauma.

Dermatological examination showed exophytic, painless, ulcerated tumor with sanguinolent exudation, measuring 10x5 cm in size, in the left parietal region (Figure 1), and several other smooth-surface tumors, soft consistency, measuring 1 to 3 cm in diameter, distributed on the scalp, some surrounded by alopecia (Figure 2). There was no palpable adenomegaly in the cervical and scalp. Two lesions were excised (Figure 3), whose pathological examination showed squamous cell proliferation in the dermis with formation of tumors with irregular multiple lobules of varying sizes, differentiating into large keratinocytes with discrete atypia, abrupt keratinization with foci of calcification and absence of invasion of adjacent tissues (Figure 4).

Trichilemmal tumor, pilomatrixoma, protuberant dermatofibrosarcoma was clinically suspected, but the correlation between clinical and histopathological findings confirmed the diagnosis of PTT.

Surgical treatment of some lesions has been proposed, with wide margin exercises and simple interrupted suture with good healing results. Due to the recurrent nature of the lesions, the patient remains in outpatient clinical follow-up and other injuries are scheduled.

DISCUSSION

Trichilemmal cysts correspond to 20% of cutaneous cysts, the others classified as epidermoid cysts originating from trichilemmal or external root sheath of the hair follicle.[1,11] PTT seems to originate from these lesions, presumably after inflammation or trauma, due to the fact that have areas with benign characteristics and others with malignant properties.[1,5-12] It differs from the trichilemmal cyst because it is uncommon, larger, and histologically more atypical, and the malignant proliferative trichilemmal tumor is less aggressive.[9] PTT occurs more frequently in women (79,5-87% of cases)

Figure 1: Exophytic, ulcerated, 10x5 cm tumor with sanguinolent exudation on the left parietal region

Figure 2: Soft, smooth, some ulcerated tumors, 1 to 3 cm, surrounded by an area of alopecia on the scalp

Figura 3: Extensive excision of two proliferating trichilemmal tumors on the scalp

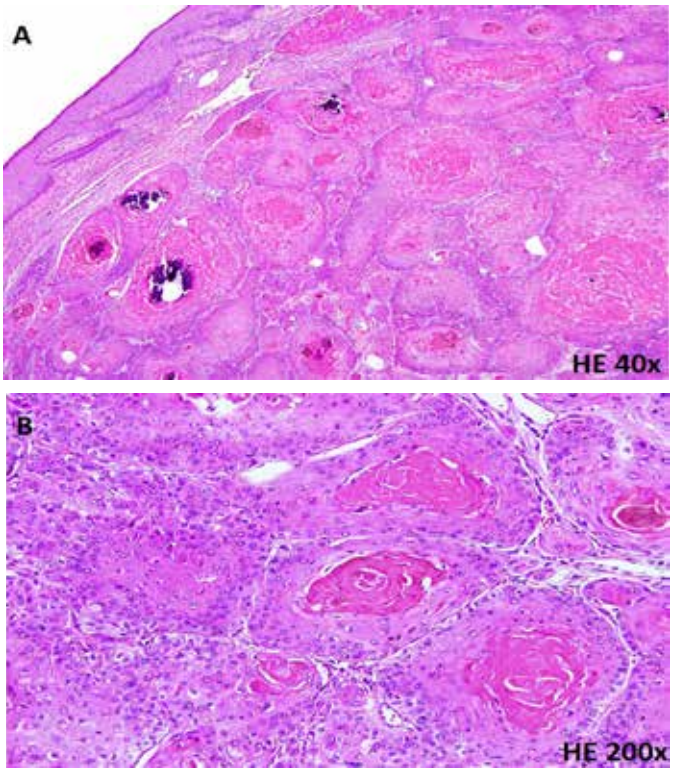

Figure 4: (A) Dermal tumor with multiple lobules varying in sizes, squamous cell proliferation with abrupt keratinization and foci of calcification. **(B)** Squamous epithelial lobules with discrete atypia associated with amorphous keratin mass and absence of inflammatory infiltrate

with ages ranging from 21 to 88 years, mean 62.4 years, despite reports in children under 18 years.[1,3,5,10-12] Lesions are usually solitary nodules, smaller than 1 cm to 12 cm (average 3.3 cm), although there are citations of multiple nodules, sometimes ulcerated and larger than 25 cm.[3,10-13] It often presents a history of slow and progressive growth from months to years, followed by fast growth and exophytic appearance.[2,10,12] The preferred location is the scalp (85.4-90% of cases) and sometimes has alopecia.[1,3,5,10-13] The remaining 10% are mostly on the back and more rarely elsewhere, such as frontal, nose, eyelids, lips, oral cavity, neck, trunk, genital, buttocks, upper and lower limbs, and skull base.[1,3,5,10-12] The reported case manifested unusual features, such as the onset of PTT at 17 years, the presence of multiple nodules and some ulcerated lesions. Most of the PTT presents benign biological behavior, but with a chance of recurrence, in a period of six months to more than 10 years after surgery, especially in cases of cleft excision , and in very rare cases may have malignant biological potential.[3, 5,10-12] Local recurrence rates (3.7%) and lymph node metastasis (1.2%) are low, but when occur they may cause invasion into deep tissue planes by continuity, contiguity or spread, resulting in high morbidity rates and even mortality.[3,5,10-12] Some studies have shown the importance of researching Ki-67 immunoexpression, a protein responsible for cell proliferation, because the expression index correlates with the degree of malignancy of the tumor.[7,8] The differential diagnosis of PTT includes squamous cell carcinoma, basal cell carcinoma, keratoacanthoma, pilomatrixoma, sweat gland tumor, protuberant dermatofibrosarcoma, cylindroma, epidermoid cyst, malignant PTT, and angiosarcoma.[1,4,11] Histopathological findings of squamous epithelium lobules, cells with abundant eosinophilic cytoplasm with abrupt keratinization, variable cytological atypia and absence of infiltration into adjacent stroma.[1,4,12,13] PTTs are recurrent and potentially invasive, so, the therapy of choice is surgeries with a margins of 0.5 to 1 cm.[5,7,9-12] Among surgical techniques, Mohs' micrographic surgery has a good indication, because its margins may to exceed visible clinical limits and thus reduce the risk of relapse and metastasis.[5,9,11] Other treatments such as lymph node dissection, radiotherapy, and chemotherapy may be required in cases of metastatic dissemination or transformation to malignant PTT.[5,11] Therefore, long-term follow-up is indispensable.[7,10-12]

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